

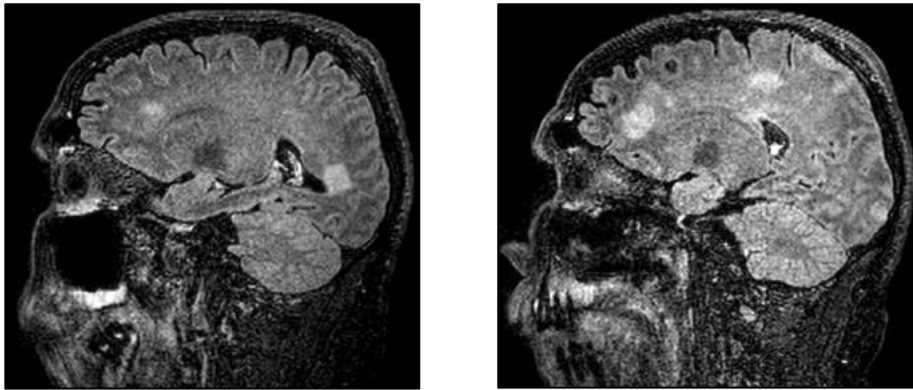
# CNS Intravascular Large B-Cell Lymphoma mimicking acute neurovasculitis: case report with autopsy and review of the literature

Leticia Ussem\*; Livia M. Thomazini; Jamile B. Oliveira; Caio M. Nagara; Fabiano P. Saggiaro; Fernando Chahud; Simone G. Ramos; Luciano Neder\*\*  
Dept. of Pathology and Forensic Medicine, Faculty of Medicine of Ribeirão Preto, University of São Paulo (FMRP-USP)

\*leticiussem@gmail.com; \*\*neder@fmrp.usp.br

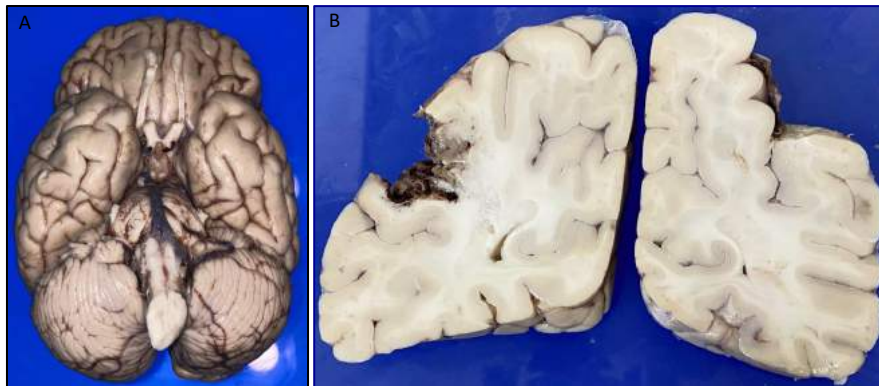
## Case Presentation

A 55-year-old male was admitted to the hospital due to unprecedented seizures. Except for vertigo and mild cognitive impairment, his previous clinical history was unremarkable.



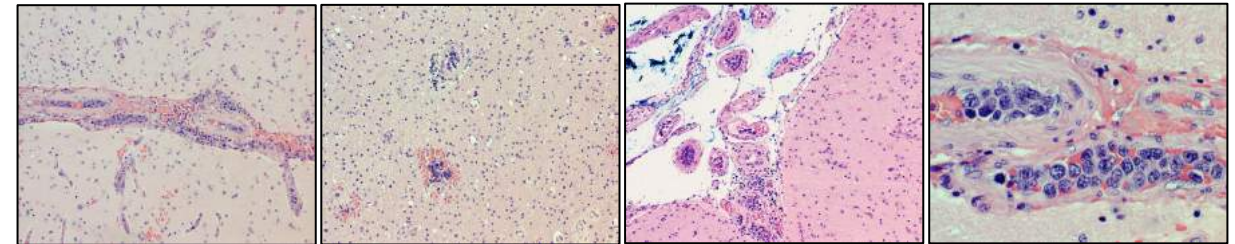
**Fig. 1.** FLAIR MR image disclosed signal abnormalities in periventricular areas.

Unfortunately, the patient expired few hours after the biopsy due to extubating failure. Necropsy was requested.

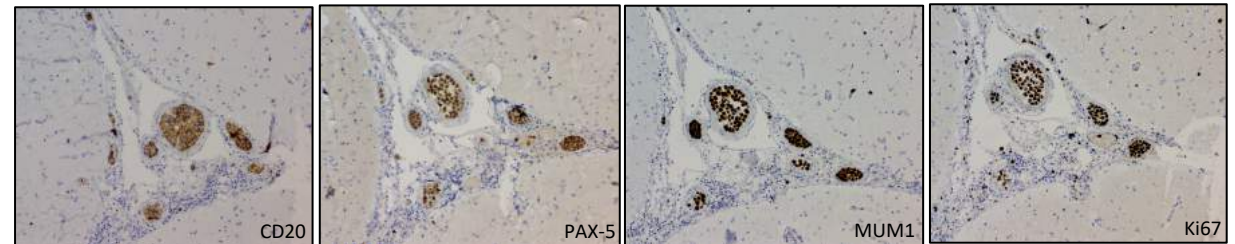


**Fig. 2.** At neuropathology examination, brain edema and irregular white matter lesions at periventricular region associated with watershed cortical infarctions were observed, besides the area from the previous biopsy (Fig. 2B).

References:  
Wu M, Lin Y, Huang X, Zhang B. Intravascular large B-cell lymphoma presenting as rapidly progressive dementia and stroke: A case report. *Medicine (Baltimore)*. 2021; 100(48):e27996 [doi: 10.1097/MD.00000000000027996].  
Belli E, Milano C, Pesaresi I, Trivelli I, Tavoni A, Ciancia E, Ali G, Zampa V, Pizzanelli C, Siciliano G, Ricci G. A case of intravascular large B cell lymphoma with brain involvement mimicking progressive multifocal leukoencephalopathy. *Int J Neurosci*. 2021; 1-5. [doi: 10.1080/00207454.2021.1972418].  
Turin CG, Ting K, Bradshaw A, Dunham SR, Nunez-Wallace K, Patel SM, Dangayach P, Holdener S, Lin WC. Central nervous system intravascular lymphoma leading to rapidly progressive dementia. *Proc (Bayl Univ Med Cent)* 2021; 34(3):373-375 [doi: 10.1080/08998280.2020.1866943].  
Liu Z, Zhang Y, Zhu Y, Zhang W. Prognosis of Intravascular Large B Cell Lymphoma (IVLBCL): Analysis of 182 Patients from Global Case Series. *Cancer Manag Res* 2020; 12:10531-10540 [doi: 10.2147/CMAR.S267825].



**Fig. 3.** Histological examination of the surgical specimen showed diffuse brain parenchyma edema and few inflammatory cells. seen aggregates of pleomorphic and large sized cells confined to cerebral blood vessel lumen.



**Fig. 4.** These cells showed strong IHC expression of CD20, PAX5 and MUM1. The Ki-67 labeling index was > 95%. The final diagnosis was CNS intravascular large B-cell lymphoma, non-germinal center type (CNS-IVLBCL).



**Fig. 5.** Interestingly, IVLBCL was just found in the biopsy area, despite extensive brain sampling.

## Discussion

CNS-IVLBCL is a very rare type of cerebral lymphoma with few autopsy reports. It is postulated that the absence of CD29 and CD54 (ICAM) by tumor cells underlie their inability to migrate transvascularily. The median patients' age is 70 years (34 – 90 years), with no sex predilection. Prognosis is poor as life expectancy after diagnosis is < 1 year. The differential diagnosis with encephalitis and vasculitis can be difficult because the overlapping of clinical and radiologic features. The gold standard for diagnosis is the brain biopsy, excluding neuroinfections, vasculitis or demyelination diseases, as seen in the current case.